HEMATOLOGY WAIVERS

CONDITION: ANEMIAS/CONGENITAL (INCLUDING SICKLE CELL TRAIT AND THALASSEMIA TRAIT)

Revised March 2003

AEROMEDICAL CONCERNS: Anemia is defined as a decrease in the hemoglobin concentration of whole blood below the lower limit of the normal range. It varies with age and sex. Anemia may arise acutely following blood loss, from decreased red cell production, or from increased destruction (hemolysis), and may be acquired or congenital. The degree of anemia and resultant end organ hypoxia, as well as the underlying disorder that may have been responsible for the anemia both represent major concerns to aviation safety. While most individuals are asymptomatic with whole blood hemoglobin concentrations above 10.0 g./dl. or hematocrits above 30 percent, the clinical manifestations of anemia will vary with the speed of onset and the physical activity of the patient. While there is little data available relating the degree of anemia to aviation activity, individuals with hemoglobin levels less than 12.0 g./dl., and 11.0 g./dl., for men and women respectively, are not eligible for flying duty.

Congenital anemias of concern in the adult include thalassemias, hemoglobinopathies, and other hemolytic anemias. The thalassemias are characterized by the deficient production of one of the globin chains of the hemoglobin molecule. According to currently used nomenclature, the disease is defined by the chain that is deficient and the number of missing chains. Beta-thalassemia denotes deficient production of beta chains. Absence of one of the two chains usually results in a subclinical condition termed beta thalassemia trait. Similarly, deficient production of alpha chains defines alpha-thalassemia. Absence of one or two of the four alpha chains results in a subclinical (alpha thalassemia trait) or mild hemolytic anemia, respectively. Loss of both beta chains or more than two alpha chains results in a severe microcytic, hypochromic anemia that is disqualifying for military duty.

Hemoglobinopathies result from the structural alteration of one the globin chains, most commonly due to single amino acid substitutions that alter the solubility, stability, or function of hemoglobin. Hemoglobin S alters the solubility of hemoglobin causing it to crystallize at low oxygen tensions. Patients with sickle cell disorders (hemoglobin SS – sickle cell anemia, hemoglobin SC disease, and hemoglobin S – thalassemia) develop a normochromic, normocytic hemolytic anemia. They are at markedly increased risk for vaso-occlusive episodes, including infarction, involving the spleen, lungs, brain, and kidneys, when exposed to hypoxia, infection, dehydration, or cold. Cases of painful crises have been reported at altitudes as low as 2,500 feet. Individuals with Sickle Cell Trait (inherit only one hemoglobin S gene) are usually asymptomatic, although they are at a slightly increased risk for intravascular sickling at altitudes above 21,000 feet.

Other congenital hemolytic anemias are due to membrane structural defects, including hereditary elliptocytosis and spherocytosis, and red cell enzyme deficiencies, including glucose 6-phosphate dehydrogenase deficiency. Hereditary elliptocytosis is a common condition and usually is subclinical or results in only very mild anemia. Hereditary spherocytosis, on the other hand, may cause a wide spectrum of disease, from mild to severe hemolysis. Glucose 6-phosphate dehydrogenase deficiency is an X-linked disorder affecting males. Severity of the disorder varies markedly according to the mutation involved and the oxidant stress placed on the red cells. The variety affecting African-Americans usually causes mild compensated hemolysis with only mild anemia. These individuals are, however, susceptible to the occurrence of severe hemolysis after the ingestion of oxidant drugs. The antimalarials are particularly hazardous.

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WAIVERS: Initial Applicants (All Classes) and Rated Aviation Personnel (All Classes): Congenital Hemolytic Anemias, including Thalassemia and Sickle Cell Disease, are disqualifying for all aviation duty. Individuals with thalassemia trait and sickle cell trait are usually granted an Exception to Policy or Waiver as long as the hemoglobin level is not less than 12.0 or 11.0 g./dl. for men and women respectively, and in the case of sickle cell trait, there is no history of vaso-occlusive crises. Any occurrence of vaso-occlusive crisis on exposure to altitude in flight or in the decompression chamber is disqualifying for all flying duty. Congenital conditions that do not result in anemia (eg. Elliptocytosis, hemoglobinopathy traits) and are asymptomatic may be classified as Information Only.

INFORMATION REQUIRED: The following clinical and laboratory data is required for an exception to policy or waiver request:

1. Clinical history of the condition, including diagnosis and course.
2. CBC with reticulocyte count.
3. Hemoglobin Electrophoresis in cases of thalassemia and hemoglobinopathies. (In the case of sickle cell trait, the electrophoresis must document hemoglobin A > hemoglobin S).
4. Hemoglobin A_2 quantification in cases of beta-thalassemia trait.
5. Serum iron, TIBC, and serum ferritin in cases of thalassemia trait.
6. Internal Medicine or Hematology Consultation.

FOLLOW-UP: Complete CBC with all comprehensive FDME. For individuals with sickle cell trait, annual evaluation on FDME/FDHS for any indicators of vaso-occlusive crises.

TREATMENT: N/A. (For aviators with sickle cell trait, avoidance of risk factors for intravascular sickling, as noted below).

DISCUSSION: The differential diagnosis of anemia is extensive. The local flight surgeon may proceed with the evaluation of anemia to his/her level of expertise. In general, the diagnosis of congenital anemias will have been made prior to the FDME and entail the review of clinical history and confirmatory laboratory tests. Decisions as to eligibility for flight status are determined by the waiver criteria. Interim assignment of a DNIF status should be based on the level of hemoglobin concentration, the rate of its decline, and the health of the aviator. Unexpected symptoms or a fall of hemoglobin to less than 11.0 g./dl. should result in prompt grounding and evaluation.

In addition, individuals with sickle cell trait should be counseled on risk factors for intravascular sickling, including hypoxia, and volume depletion. Finally, they should be counseled on the dangers associated with recreational diving and general anesthesia. It is generally accepted that those with hemoglobin S trait are not at significantly increased risk from general anesthesia.

REFERENCE: Merck Manual, Chapter 27 found online at:
http://www.merck.com/pubs/mmanual/